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Demographic and Clinical Characteristics of Children with Guillain-Barre Syndrome

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Abstract: ***Background:*** Guillain-Barre Syndrome (GBS) is a rare neurological disorder affecting all age groups, characterized by the immune system mistakenly attacking peripheral nerves, causing weakness, numbness, and, in severe cases, paralysis. While the exact cause is often unknown, GBS is commonly preceded by infections like respiratory or gastrointestinal illnesses. ***Aim of the study:*** This study aimed to assess the demographic and clinical characteristics of children with Guillain-Barre syndrome. ***Methods and Materials:*** This observational study was carried out at the National Institute of Neurosciences and Hospital (NINS&H) in Dhaka, Bangladesh, spanning from September 2018 to August 2019. A total of 93 children diagnosed with childhood Guillain-Barre Syndrome (GBS) were included as the study subjects using a consecutive sampling technique. Data analysis was performed using MS Office tools. ***Results:*** Participants, with a mean age of 7.73 ± 3.80 years and a male-to-female ratio of 1.7:1, reported antecedent illnesses in 60 cases (64.5%). Electrophysiological abnormalities indicated 65.6% with acute motor axonal neuropathy (AMAN), 31.2% with acute inflammatory demyelinating polyradiculo-neuropathy (AIDP), and 3% with acute motor and sensory axonal neuropathy (AMSAN). Neurologically, 87.1% exhibited progressive weakness (quadriparesis), while 12.9% had paraparesis. Cranial nerve dysfunction, including 42% with bulbar involvement and 2.1% with facial palsy, was observed in 44.1% of cases. ***Conclusion:*** Guillain-Barré Syndrome (GBS) tends to affect younger male children more frequently.

Keywords: Demographic characteristics, Clinical, Children, Guillain-Barre syndrome, GBS, Quadriplegia.

Original Research Article

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Article at a glance:

Study Purpose: *To assess the demographic and clinical characteristics of children with Guillain-Barre Syndrome (GBS) in Dhaka, Bangladesh.*
Key findings: *The study found that GBS in children is commonly preceded by infections, and a variety of clinical features such as weakness, numbness, and paralysis were observed.*
Newer findings: *The research provides regional data on the clinical presentation of GBS in children, which could help refine diagnostic and treatment protocols in Bangladesh and similar settings.*
Abbreviations: *GBS – Guillain-Barré Syndrome, NINS&H – National Institute of Neurosciences & Hospital, PEDS – Pediatrics, MS Office – Microsoft Office, RCT – Randomized Controlled Trial.*

INTRODUCTION

Guillain-Barré syndrome (GBS) encompasses a collection of clinical syndromes characterized by acute polyneuropathy resulting from an immune-mediated process. Typically, individuals with GBS exhibit progressive weakness that may affect the autonomic, bulbar, and respiratory systems, accompanied by diminished

or absent deep tendon reflexes. [1] With the eradication of polio, GBS has emerged as the most common cause of acute or sub-acute flaccid weakness. The reported annual incidence rates of GBS in children range from 0.34 to 1.34 per 100,000.[2,3] GBS is diagnosed clinically when there is a presence of progressive limb weakness along with areflexia or hyporeflexia [4]. While Guillain-Barré syndrome (GBS) can manifest at any age, it is notably prevalent in children between the ages of 1 and 5 years.[5, 6] Although there have been documented instances of GBS in newborns, its occurrence in infants is rare. [7]

The syndrome exhibits a slight male predominance, with a male-to-female ratio of 1.5:1. While most GBS cases are sporadic, there have been instances of epidemic clusters. In northern China, the motor axonal form of GBS tends to occur in epidemics during the summer, predominantly affecting children, possibly due to campylobacter jejuni infection [8]. Serological evidence of recent campylobacter jejuni infection was discovered in approximately half of the children with GBS in the same region. [9] Supporting features for the diagnosis of Guillain-Barré syndrome (GBS) include a progression of symptoms lasting up to 4 weeks, relative symmetry of weakness and sensory loss, sensory symptoms being less prominent than weakness if present, pain in the back and legs, autonomic dysfunction, absence of fever, albuminocytologic dissociation in cerebrospinal fluid (CSF) studies, and post-gadolinium enhancement of peripheral nerve roots and cauda equina in magnetic resonance imaging (MRI).[10] Acute inflammatory demyelinating polyneuropathy (AIDP), which affects segments of the myelin sheath, is the most common clinical subtype of GBS. There are also fewer common subtypes, including acute motor axonal neuropathy (AMAN) and acute motor and sensory axonal neuropathy (AMSAN), which target neuronal axons, and Miller-Fischer syndrome, characterized by ataxia, areflexia, and ophthalmoplegia.[11] The presenting symptoms of Guillain-Barré syndrome (GBS), such as flaccid paralysis and areflexia, overlap with a wide range of conditions affecting the cerebellum, spinal cord, peripheral nerves, and muscles. This often necessitates extensive and costly medical investigations for differential diagnosis [4]. In comparison to adults, children generally have a better prognosis, with approximately 80% of children experiencing full recovery without long-term recurrence.[4] However, a small subset of children may have residual symptoms, necessitating lengthy and costly rehabilitation.

METHODS

The observational study was carried out at the National Institute of Neurosciences and Hospital (NINS&H), Dhaka, Bangladesh, spanning from September 2018 to August 2019. The study included a total of 93 cases under the age of 16, who were admitted to the Department of Pediatric Neurology of the mentioned hospital with childhood Guillain-Barré Syndrome (GBS). The cases were also selected from direct admission to ICU and HDU because of impending respiratory failure utilizing a consecutive sampling technique. The study received approval from the ethical committee of the mentioned hospital, and written consent was obtained from all participants before data collection. The inclusion criteria comprised children and adolescents below 16 years diagnosed with Guillain-Barré Syndrome (GBS) based on the Asbury and Cornblath criteria. Conversely, patients with GBS and coexisting conditions such as cerebral palsy, hereditary neuropathy, and congenital myopathy were excluded based on the study's exclusion criteria. Demographic and clinical information for all participants was meticulously recorded, and data analysis was conducted using MS Office tools.

RESULT

In this study, the participants had a mean age of 7.73 ±3.80 years, with the highest number (41.9%) falling in the 5-10 years age group, followed by 31.2% in the 1-5 years age group. There was a male preponderance, with a male-to-female ratio of 1.7:1. Antecedent illnesses were reported in 60 (64.5%) children, with 26.9% having a diarrheal episode, and 20.4% experiencing respiratory tract infections. Additionally, 17.2% had a history of fever or nonspecific symptoms, while 35.5% had no preceding symptoms. The study found no significant seasonal variation, with 34.4% of cases occurring in winter, followed by 33.3% in summer and 32.3% in the rainy season. The majority of patients experienced a time-to-peak deficit between 3 and 5 days, with a mean time of 5.37 ± 2.8 days.

The study identified various subtypes of Guillain-Barré Syndrome (GBS) based on electrophysiological abnormalities. Among the participants, 65.6% exhibited acute motor axonal neuropathy (AMAN), 31.2% presented with acute inflammatory demyelinating polyradiculo-neuropathy (AIDP), and 3% showed acute motor and sensory axonal neuropathy (AMSAN). Notably, axonal types (AMAN and AMSAN) of GBS were more prevalent, accounting for 68.8%, compared to the demyelinating type, which represented 31.2% of cases. In the neurological presentation analysis, the majority of patients (87.1%) exhibited progressive weakness of all four limbs (quadriparesis), while 12.9% presented with paraparesis. Cranial nerve dysfunction was observed in 44.1% of cases, including bulbar involvement in 42% and facial palsy in 2.1%. Other clinical features included pain (58.1%), nasal intonation (23.7%), neck flexor weakness (22.6%), breathing difficulty (22.6%), and autonomic dysfunction (7.5%). MRC scoring, possible in 64 patients, revealed that 67.2% had a score between 21 and 40 (Moderate weakness) at admission. Most patients with a baseline HMS score of G-4 were wheelchair-bound at admission.

Table 1: Age distribution of subjects (N=93)

Age (Years)	n	%
<5	29	31.2%
5-10	39	41.9%
>10	25	26.9%
Mean ± SD	7.73±3.80	

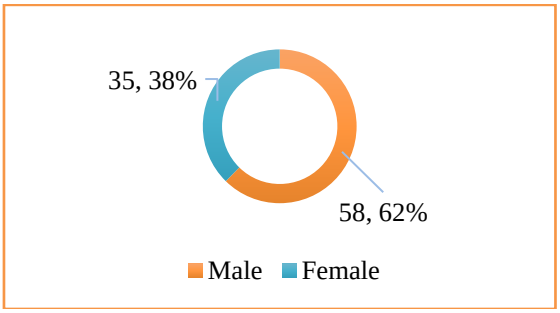

Figure 1: Gender distribution

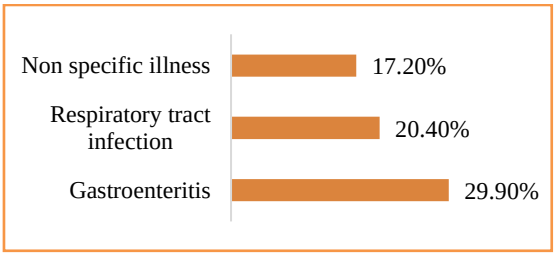

Figure 2: Antecedent event distribution

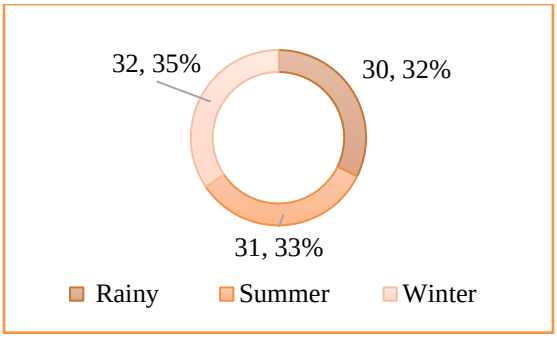

Figure 3: Distribution of seasonal variation

Table 2: Time to peak deficit

Days	n	%
<3	27	29.0%
3-5	33	35.5%
6-10	21	22.6%
>10	12	12.9%
Mean ± SD	5.37±2.8	

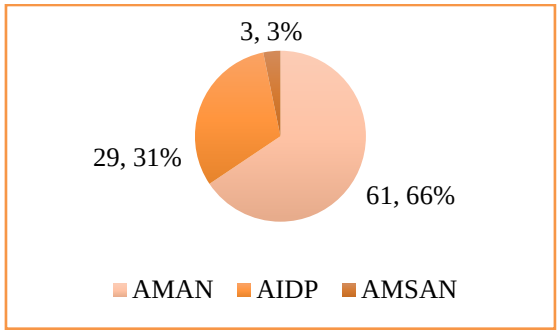

Figure 4: Varieties of GBS based on electrophysiological pattern

Table 3: Neurological presentation

Parameters	n	%
Variables		
Quadriplegia	81	87.10%
Paraplegia	12	12.90%
Cranial nerve involvement	41	44.10%
Bulbar involvement	39	41.90%
Facial palsy	2	2.10%
Breathing difficulty	21	22.60%
Nasal intonation	22	23.70%
Autonomic involvement	7	7.50%
Pain	54	58.10%
Neck flexor weakness	21	22.60%
Baseline MRC sum score (n=64)		
Severe weakness (0-20)	13	20.30%
Moderate weakness (21-40)	43	67.20%
Mild weakness (41-60)	8	12.50%
Baseline HMS score (n=93)		
Walk 5-meter support (G-3)	10	10.80%
Wheelchair-bound (G-4)	65	69.90%
Ventilator support (G-5)	18	19.40%
(Mean ± SD)	4.09 ± 0.54	

DISCUSSION

This study aimed to assess the demographic and clinical characteristics of children with Guillain-Barre syndrome. In the current study, the majority of patients (41.9%) were aged 5-10 years, with a mean age of 7.73 ± 3.80 years. The youngest patient was a 15-month-old male child, and the oldest was 14 years old. Comparatively, Koul et al. reported most children below 4 years of age in their study of 61 children under 15 years, while Kumbhar et al. found that 50% of their patients presented between 6 months to 5 years of age [12, 13]. In the current study, males were more prevalent, accounting for 58 (62.4%), with a male-to-female ratio of 1.7:1. This finding aligns with the observation of male preponderance in the study by

Dhadke et al., where the male-to-female ratio was 1.5:1 [14]. Similar results were reported in a Kolkata study conducted by Sarkar et al., where 69.06% were male [15]. The similarity in gender distribution across these studies may be attributed to shared sociocultural conditions and the cultural emphasis on male children in this subcontinent. In our study, two-thirds (64.5%) of participants had a history of antecedent illnesses such as diarrhea and acute respiratory infections (ARI). Similar findings were reported by Kumbhar et al. (69.12%) in a study conducted in India [13]. Winner Hughes et al. also observed that more than 50% of cases had a significant history of prior illness [16].

Gastroenteritis was the most common antecedent illness in our study, affecting 25 (26.9%) individuals, followed by respiratory tract infection in 19 (20.4%), although the specific organisms were not identified. In contrast, Kumbhar et al. found respiratory tract infections to be more prevalent in their study, accounting for 38.4% of cases [13]. In the current study, nerve conduction studies (NCS) were conducted in all patients, revealing that acute motor axonal neuropathy (AMAN) (66%) was the most common variant of Guillain-Barré Syndrome (GBS), followed by acute inflammatory demyelinating polyneuropathy (AIDP) (31%) and acute motor and sensory axonal neuropathy (AMSAN) (3%). Verma et al. also reported that two-thirds of their patients exhibited the axonal pattern of GBS in their study [17]. In contrast, AIDP was found in 63% of cases in a study conducted by Barzegar et al. [18]. This finding contrasts with the usual predominance of AIDP over the axonal form of GBS, especially in Western countries [19]. Similar results were observed in the study conducted by Ropper et al., where acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), and acute motor and sensory axonal neuropathy (AMSAN) were present in 63%, 23%, and 14% of patients, respectively [20]. However, the AMAN variety is now considered the most common form of Guillain-Barré Syndrome (GBS) in Asian countries [21].

Symmetrical muscular weakness with areflexia or hyporeflexia is observed in 90% of Guillain-Barré Syndrome (GBS) cases, as reported by various studies, including Haden et al [22]. Our study also showed a similar pattern, with 87.1% of patients presenting with flaccid quadriparesis and 44.1% experiencing cranial nerve involvement. This aligns with findings from the study conducted by Habib R et al, where 84% of patients exhibited quadriparesis and 48% had cranial nerve palsy [23]. The acute phase of GBS can lead to life-threatening complications, including respiratory paralysis, secondary infections, bulbar weakness, multisystem involvement, and dysautonomia [24,25]. Islam Z et al highlighted that respiratory failure and oropharyngeal weakness may necessitate ventilator assistance in approximately one-third of hospitalized patients, underscoring the critical importance of early management in Guillain-Barré Syndrome (GBS) [26]. Veena Kalra et al reported a high rate of ventilation (19.2%) and acute phase mortality (11.5%) in their study conducted in India [27]. Acute respiratory failure was also noted in 20% of cases in the study by Haden et al [22]. Similar findings were observed in studies conducted by Koul et al (17.3%) and Kumbhar et al (19.23%) [12, 13]. A study by Rees in South East England found that 25% of their patients required mechanical ventilation [25]. In our study, acute respiratory failure was observed in 18 (19.4%) patients who required mechanical ventilation, and there were no reported mortalities, consistent with the findings of the study by Koul et al. [12].

Both bulbar and neck flexor weakness are commonly associated with respiratory compromise, likely because oral and pharyngeal protective reflexes are impaired in such patients. Neck flexor weakness often correlates with diaphragmatic weakness, leading to the need for mechanical ventilation. Rajesh et al. and Paul et al. also reported a high prevalence of bulbar involvement, with 92.5% and 86.6% of patients, respectively, although our study indicated that 42% of patients had bulbar involvement [17,28]. In the study by Rajesh et al., autonomic dysfunction was demonstrated in 34.4% of adults with GBS, while our study showed this in 9 (9.7%) patients.[17-36] The relatively lower proportion of patients with autonomic dysfunction in our study may be attributed to the fact that the evaluation for such abnormalities relied solely on symptoms and signs, without specific testing for autonomic dysfunction, or it could be related to geographic variation or other genetic factors. Meticulous respiratory care,

prompt management of secondary infections, and monitoring for bulbar weakness and dysautonomia, along with appropriate supportive care, may contribute to the favorable outcomes observed in our study.

Limitation of the study:

The study is limited by several factors. Firstly, the small sample size restricts the generalizability of findings. Secondly, as a single-center study, the results may lack diversity. Lastly, being conducted in a tertiary care hospital, the prevalence may be skewed, making it challenging to extrapolate findings to the broader population. Future population-based studies are imperative for a more comprehensive understanding.

CONCLUSION & RECOMMENDATION

Guillain-Barré Syndrome (GBS) demonstrates certain demographic and clinical patterns in affected children. It tends to more frequently affect younger male children. Unlike some diseases, GBS doesn't exhibit a notable seasonal variation. A characteristic feature of GBS in children is the presence of progressive weakness. Recognizing these patterns is crucial for timely diagnosis and appropriate management in pediatric patients affected by Guillain-Barré Syndrome.

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